Lower limb lengthening is not indicated in familial short stature

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Auxology is the study of growth (the increase in height, weight, and head circumference, in childhood and adolescence) (Medical Dictionary)

1. Introduction

Surgery for the lengthening of bones in the setting of lower limb discrepancy were highly influenced by the works of Ilizarov and the discovery of callotasis. Use of this technique in order to achieve an overall increased length for cosmetic purposes is rapidly gaining popularity.

The invasive nature of this surgical procedure requiring an external fixator has set forth an ethical dilemma, and only a few surgeons have accepted the idea of undertaking lengthening procedures of both limbs for this purpose.

The constant progress in the realm of lengthening surgeries and the advent of motorized nails have undoubtedly allowed the pondering of such a dilemma on a more acceptable risk to benefit ratio.

Pediatric orthopedic surgery departments are being increasingly consulted with such demands by patients who find themselves shorter than their peers. How could a surgeon react when faced with such a demand?

2. Definition of short stature

It is difficult to define with certainty the threshold separating normal from pathologic height. In terms of growth, a “normal” stature may be defined as the height attained by the majority of the healthy population.

Sempé and Pédron’s growth charts for the French population [1] show an average height of 1.63m [1.52m – 1.74m] for girls, and 1.75m [1.63m – 1.87m] for boys. Short stature is not

necessarily pathological, since hereditary factors intervene significantly in the determination of height [2]. Nevertheless, syndromic etiologies do exist and may reveal an underlying pathology. As such, there are many diseases that may lead to short stature. Included among these pathologies are those of the larger group of skeletal dysplasias with or without involvement of the growth plates, or endocrine, nutritional, and genetic causes. Thus, the growth of a child should be followed closely and regularly, thus allowing to screen early on for growth disturbances. For a certain number of subjects presenting a secondary small stature, medical treatment by growth hormone may be considered during childhood, thus modifying the prognosis of the expected final height. By the end of growth, skeletal lengthening may be achieved only by surgical means.

3. Bone lengthening: History

Although the first records of bone lengthening seem to emanate from the XVIth century by Saint-Ignac of Loyola, it is not until 1904 that the first publication appeared on successful bone lengthening by Codivilla [3]. The first external fixators were detailed in 1912 by Ombreddane [3]. In 1971, Wagner popularized extemporaneous lengthening of bones using an external fixator followed by osteosynthesis with a bone graft [4] (fig 1).

The first revolution of these lengthening techniques came from Ilizarov's works in Siberia [5,6]. Ilizarov introduced two major concepts in lengthening: a stable lengthening apparatus and the biological concept of bone regeneration in distraction. In 1987, G. de Bastiani improved the callotasis technique [7,8] by associating Ilizarov's principles with a unilateral external fixator (fig 2). The high rate of complications related to bone lengthening on external fixators made the indication debatable. In 1989, the first censused publication reported the case of three patients with achondroplasia [9].

The second revolution came from Taylor in 1991, who developed a hexapodal circular external fixator allowing progressive three-dimensional computer-guided correction [10]. As a result, this technological advancement allowed the correction of bony deformities simultaneously with lengthening. This technique has been useful in patients with pathological short stature since bony deformities are often associated (fig 1).

In order to address the numerous complications associated with external fixators, numerous authors proposed the adjunction of centromedullary osteosynthesis by a Kirschner wire. Popkov's works showed a decrease in the healing index and decreased axial complications.

Finally, the third major evolution took place in the 1990s with the advent of progressive bone lengthening on a singular centromedullary nail [12], allowing to considerably reduce the number of complications related to bone lengthening with external fixators [13] (fig 2).

Since lengthening was becoming increasingly less invasive, a final step was needed to propose this lengthening technique to be used for cosmetic reasons to see the light of day. The advent of

motorized nails also further modified the point of view of certain surgeons thus rendering the idea of lengthening surgeries solely for cosmetic purposes more acceptable.

4. Summary of a meta-analysis

In 2020, Marwan et al. [14] published a meta-analysis on this subject. Based on the PubMed and Embrase databases, 11 studies on 795 patients were included. The M:F ratio was found to be 1.6:1, mean age at surgery was 26 years and 1 month old. Mean height of patients before surgical intervention was 160cm [130cm – 180cm]. Ilizarov-type external fixators with or without centromedullary K-wires was the most utilized technique. Lengthening nails represented only 8% of patients. Most surgical interventions were undergone on the tibia, 3 out of the 11 studies privileged femoral lengthening, and 2 studies out of 11 utilized a mixed femur/tibia technique. A minority of patients (3.8%) also underwent simultaneous axis correction.

For patients who underwent external fixator lengthening, the average duration of fixator use was of 201 days [24 days – 810 days], the index of consolidation was of 29.2 days/cm [6.3 days/cm – 180 days/cm]. The average follow-up was of 4.9 years [41 days – 7 years]. Most patients were satisfied of the results and reported excellent function. Nevertheless, many complications were reported in almost all patients with a mean 0.78 minor complications, 0.94 moderate complications, and 0.15 major complications per patient. The gain in length was more important in the external fixator/nailing group. Patients who underwent lengthening on a nail had fewer complications. Thus, the authors concluded that during lengthening for small stature:

- Gain in length is substantial with excellent satisfaction from patients, and most patients reported great satisfaction following their intervention and recommended the same intervention to other patients.
- Although familial short stature might not be considered pathological, it may lead to psychological and functional disorders, and may impact quality of life.
- Short stature might be more stigmatizing and more difficult to accept for men than women.
- Lengthening procedures are responsible for a number of complications that are relatively severe and non-negligeable (especially when this surgery is undergone using an external fixator).
- Bone lengthening with an external fixator is a relatively long process.
- Conversely, there is a net decrease in complications and duration of treatment when the lengthening is undergone using a centromedullary nail.
- A psychological assessment is recommended pre-operatively. In fact, patients with disorders such as dysmorphophobia would not benefit from cosmetic lengthening.

5. Management and cost of the intervention

The surgical code (NDAA003) for the progressive lengthening of the femur or the tibia by internal fixation without tendon lengthening is not covered by the social security. Nevertheless, bilateral

lengthening of the lower limbs may only be billed on case of achondroplasia or dwarfism. Concerning unilateral lengthening, the procedure may be bill if there is lower limb discrepancy >3cm and the operator is a competent orthopedic and/or pediatric surgery. It must be undergone in a specialized center with an experienced team.
As a result, the costs of lengthening procedures for cosmetic purposes would be covered by the patient). For information, the average cost of bilateral lengthening at the Rennes university hospital is of €33,298. This includes an anesthesia consultation of €25, over fifteen pediatric orthopedic consultations worth a total of €345, radiographic examinations worth €1,000, 6 days of hospitalization costing €5,532, and implantable medical devices (€12,510 + TAX at 5.5%, leading to €26,396).

6. Conclusion and response to the assertion “Lower limb lengthening is not indicated in familial short stature”

The first bone lengthening procedures were undergone on external fixators. However, this technique includes numerous complications. As a result, operating patients with this technique for cosmetic reasons is controversial. As mentioned previously, some authors took the risk and, despite the physical complications, the psychological benefits seemed certain.

Progress in terms of external fixators allowed multiplanar correction along with bony lengthening. The medical benefits in this case would render the lengthening procedure more acceptable. The advent of motorized nails also allowed a significant decrease in the number of complications. This technological advancement has allowed the risk/benefit ratio of such interventions to balance out and to reignite the debate on cosmetic bone lengthening. Nevertheless, the cost of such an intervention may lead some patients to refrain from proceeding.

The affirmation “Lower limb lengthening is not indicated in familial short stature” seems no longer justified. The demands of patients should therefore be heard and even addressed.

Figures

Fig 1: Bilateral tibia lengthening of 6cm associated with an axis correction with a hexapodal external fixator in the setting of hypophosphatemic rickets (154cm preoperative, 160cm after lengthening).

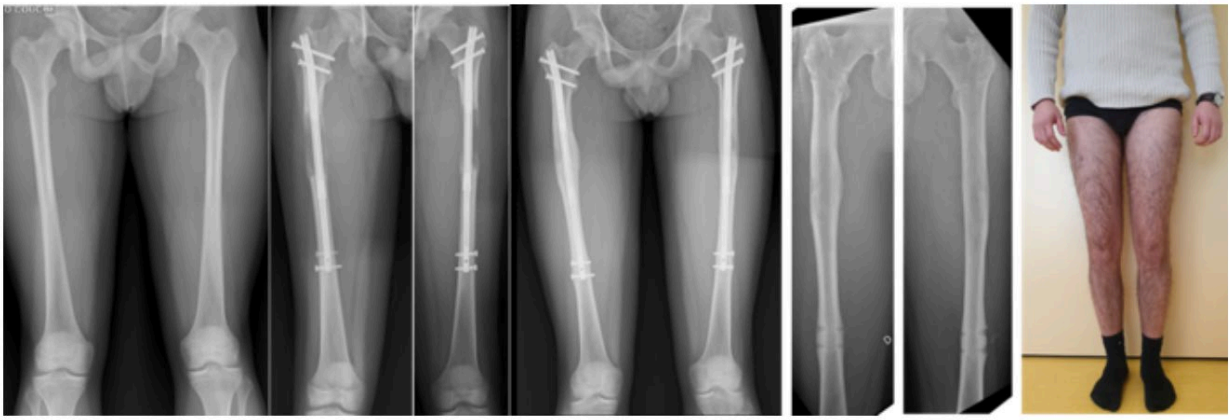

Fig 2: Cosmetic bilateral femoral lengthening of 8cm using the Precice® electromagnetic nail (163cm preoperative, 171cm after lengthening).

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